CASE REPORT

Unusual trifurcation of a single left coronary artery

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SUMMARY

Congenital coronary artery anomalies are rare and usually an incidental finding during coronary angiography. Most of the anomalies are benign in nature; however, some are malignant and may result in sudden cardiac death. A 46-year-old woman with diabetes and hypertension underwent coronary angiography for evaluation of exertional angina, which revealed an unusual trifurcation of a single left coronary artery with an anomalous origin of the right coronary artery from the left anterior descending artery but no significant coronary narrowing. The patient was managed conservatively.

BACKGROUND

An anomalous origin of the right coronary artery (RCA) as a trifurcation of the left coronary artery is an extremely rare entity. The coronary circulation from a single coronary artery usually has little clinical significance; however, variability in anatomical origin and course can cause a malignant course, resulting in sudden cardiac death and increased risk of ischaemia.

CASE PRESENTATION

A 46-year-old woman with diabetes and hypertension for the past 6 years was evaluated for exertional angina. She had a family history of premature coronary artery disease in a first-degree relative. Her blood pressure was 152/92 mm Hg prior to treatment. Blood pressure on presentation was 134/84 mm Hg in the left arm in supine position with current antihypertensive drugs. Pulse rate was 76/min, regular in rhythm, and normal in volume and character with no radioradial or radiofemoral delay, and all pulses were palpable. Other physical examinations were normal. ECG showed mild ST-T changes in precordial leads. Echocardiography revealed mild concentric left ventricular hypertrophy, grade I diastolic dysfunction and normal ejection fraction. Exercise stress testing was inconclusive as it was prematurely terminated because of dyspnoea. The patient underwent coronary arteriogram after proper consent. Cannulation of the left main coronary artery showed its normal course, which was dividing into the left anterior descending artery (LAD) and the left circumflex artery (LCx) (figures 1, 2). However, the RCA was originating from the mid-LAD after the origin of the first major diagonal branch (figures 1, 2). On aortic root angiogram using pigtail catheter, the RCA could not be visualised from the right aortic sinus (figure 3D). All the three arteries appeared free of disease.

Figure 1 Left main trunk dividing into the left anterior descending artery (LAD), left circumflex artery and right coronary artery (RCA) arising from the mid-LAD (red arrows: RCA; (A) anteroposterior caudal view; (B) anteroposterior cranial view).

INVESTIGATIONS

To confirm the anomalous origin and to better delineate the course of artery, a 128-slice multi-detector CT (MDCT) coronary angiography was performed and revealed a single left coronary artery and the RCA originating from the mid-LAD, coursing in front of the right ventricular outflow tract and further running down into the right atrio-ventricular groove. Therefore, the left main coronary artery was functionally trifurcating into LAD, LCx and RCA (figure 3A, B, C).

DIFFERENTIAL DIAGNOSIS

In our case, exercise stress testing was terminated because of dyspnoea. It could have been angina equivalence or due to increased left ventricular end-diastolic pressure because of an underlying diastolic dysfunction and mild concentric left ventricular hypertrophy. Another possibility could have been syndrome X, where patients present with chest pain and angiogram reveals normal coronary arteries with no evidence of large-vessel involvement. Of these patients, 20% may have positive exercise stress test. These patients often do not complete the exercise test because of fatigue or mild chest discomfort as in our case. Left ventricular function usually remains normal. Endothelial dysfunction, microvascular dysfunction and spasm are believed to be responsible for angina in such patients. In some cases, there is abnormal pain perception or sensitivity resulting into chest pain. Exaggerated preoccupation to personal health or panic disorder may be responsible in such cohort, but these were ruled out as there were no behavioural or psychiatric disturbances during our patient's evaluation.

TREATMENT

Initially the patient was started with amlodipine 5 mg and hydrochlorothiazide 12.5 mg once daily. As her blood pressure was not controlled, the dosage

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Figure 2 Left main coronary artery dividing into the left anterior descending artery (LAD), left circumflex artery and right coronary artery (RCA) arising from the mid-LAD (red arrows: RCA; (A) left anterior oblique caudal view; (B) straight lateral view).

of amlodipine was escalated to 10 mg, but she developed pedal oedema. To achieve better control of blood pressure, ramipril 5 mg once daily was added. Ramipril was also giving her renoprotection along with antihypertensive effect. Also, the most important is to have a strict control of blood pressure with combination therapy rather than one or two drugs with their maximal dosage, which patients may not tolerate sometimes, as in our case. Our patient was managed conservatively with antihypertensive drugs (amlodipine 5 mg once daily, ramipril 5 mg once daily and hydrochlorothiazide 12.5 mg once daily) and metformin 500 mg twice daily.

OUTCOME AND FOLLOW-UP

She was discharged on stable condition and has been on regular follow-up since then.

DISCUSSION

Anomalous RCA usually arises from the left sinus itself or directly from the left main coronary artery or its vicinity. Anomalous origin of the RCA from LAD has rarely been reported,

with only 35–40 odd cases reported in the literature.¹ Most of these cases have been reported in structurally normal heart and rarely in few patients with tetralogy of Fallot.^{2,3} Therefore, these have been identified mostly as an incidental finding and rarely present with coronary artery disease. The incidence of anomalous RCA in congenital coronary anomalies is variable, varying from 0.04% to 0.46%.^{4,5} A single coronary artery is classified if the entire coronary system originates from a single ostium in the aorta. The RCA originating from the LAD is an extremely rare coronary artery abnormality with incidence of 0.024%.⁶ The coronary circulation arising from a single coronary ostium remains silent except when coronary artery has an interarterial course between two great vessels. Although it is considered to be a benign entity, in 1982, three cases of sudden death were described among patients with this aberration.¹ It may lead to sudden cardiac death due to malignant arrhythmias as a result of repetitive ischaemia, interarterial course between the great vessels, or acute angle made by the anomalous RCA when it turns towards the atrioventricular groove causing inferior ischaemia because of reduced flow or flap-like closure of the abnormal coronary orifice.^{7–9} The RCA arising from the LAD has been considered as an extension of the second diagonal,⁶ as a large branch of the first septal perforator,^{9,10} or originating as from the mid-portion of the LAD. After origin from the LAD, they run in right atrioventricular groove and supply the same territory like the usual RCA.¹¹ In our case, the left main coronary artery was dividing into LAD and circumflex artery, and the RCA was further arising from the mid-LAD, thereby all three arteries functionally arise from single ostium from the left sinus. Catheter-based angiogram is insufficient as it does not give any information about its course; therefore, MDCT should be done to describe the anatomy and course of coronary artery as was done in our case. It also helps to rule out any small dissection that may go unnoticed by conventional angiography or other anomalies, and mechanical compression of the anomalous RCA between the aorta and pulmonary artery, which might be related to ischaemia, and therefore scores over conventional angiography. It is usually considered benign,¹² but data from case reports so far suggest that they carry increased

Figure 3 Multidetector CT coronary angiography showing the right coronary artery (RCA) arising from the mid-left anterior descending artery coursing in front of the right ventricular outflow tract (white arrows: RCA; A,B,C). Aortic root angiogram showing absence of the RCA from its usual location (D).

Learning points

- ▶ Right coronary artery can arise from the left anterior descending artery, although extremely rare.
- ▶ Although it is considered to be a benign entity, it may cause myocardial ischaemia and sudden cardiac death because of the malignant combination of acute and chronic substrates in the form of anatomy and repetitive ischaemia.
- ▶ Other causes of ischaemia could be its interarterial course between the great vessels; acute angle made by the anomalous right coronary artery when it turns towards the atrioventricular groove causing inferior ischaemia because of reduced flow; and its extrinsic occlusion during exercise.
- ▶ Multidetector CT should be done to describe the anatomy and course of coronary artery to rule out any small dissection that may go unnoticed by conventional angiography, other anomalies, and mechanical compression of the anomalous right coronary artery between the aorta and pulmonary artery.
- ▶ Microvascular dysfunction may cause angina in the absence of atherosclerosis in perimenopausal women who have diabetes or hypertension.

risk of coronary artery disease.^{10 13} Possible reasons of angina in our case in the absence of coronary atherosclerosis may be syndrome X because of microvascular dysfunction, interarterial course, and 'coronary steal' phenomenon as a result of either compromised flow in anomalous artery or acute angle of take-off for the anomalous vessel. There is no consensus about the best management strategy, and this will depend on clinical presentation, comorbid conditions and underlying atherosclerotic burden, if any.

Contributors SKS: conception and design, acquisition of data, and analysis and interpretation of data. PA: agreement to be accountable for the article and to ensure that all questions regarding the accuracy or integrity of the article are investigated and resolved. VM: drafting the article or revising it critically for important intellectual content. RT: final approval of the version published.

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